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BY

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PEMPHIGUS, AND THE DISEASES LIABLE TO BE MISTAKEN FOR IT.

IN 1877 I was asked by a medical friend to see with him a patient in whose case he was unable to arrive at a satisfactory diagnosis. The patient was a white woman, about fifty years of age, born in Ireland, but a number of years resident in this country. She had been inoculated in Ireland with smallpox virus, and passed through a mild attack of the disease at that time, the scars of which remained as evidences of the truth of her statement. About a year before I saw her, my *confrère* had attended her in an attack of smallpox of considerable severity, being the second attack, including the one following the early inoculation, that the patient had passed through.

About ten days before I was asked to see the patient, she had been attacked by the prodromic symptoms of smallpox, followed after the usual interval by an eruption of papules, rapidly developing into pustules of large size. At the time I saw her, the entire body was covered with pustules, varying in size from a pea to a hen's egg, many of them being black from effused blood. Some of the

blebs had been ruptured, discharging their contents, and leaving an uneven, ragged, necrotic-looking base, a portion of the dermal structure being evidently destroyed. There was considerable inflammation and swelling of the skin present. The patient was much prostrated, and gave all evidences of grave systemic involvement. After a thorough examination of all the circumstances, I gave it as my opinion that the patient suffered from smallpox, in spite of the fact that she had passed through two attacks of the same disease before—one (the first) according to her own testimony, supported by the evidence of the scars remaining, and the second under the care of a thoroughly competent physician, whose experience with smallpox had been gained in an epidemic of wide extent and severity a few years before. The patient died a few days after I saw her, of exhaustion.

On March 6, 1883, I was called by my friend, Dr. R. H. P. Ellis, to a case presenting somewhat similar difficulties of diagnosis. The case was that of a robust carpenter, twenty-six years of age, who consulted Dr. Ellis some days before, with a peculiar bullous eruption upon the hands and face. In view of the fact that the patient lived in a smallpox-infected part of the city, the latter disease was, of course, first thought of; but Dr. Ellis, being solicitous of doing no injustice to his patient, even at the sacrifice of some time and convenience to himself, examined him very carefully before venturing a positive diagnosis. The examination convinced him that the case before him was not smallpox, or

else that it was a very unusual and anomalous form of that disease. He therefore kept the patient under observation, treating him expectantly. The doctor noted the frequent eruption of bullæ, appearing in crops, and varying in size from a bean to a large almond. The bullæ were tensely filled with a clear, yellowish fluid, the walls of the blebs springing up abruptly from the sound skin, which presented no traces of inflammation or swelling. In a few days, the fluid contained in the blebs was absorbed, and the roof dried into a thin, brownish scab, which soon fell off, leaving a reddened, but not exuding surface beneath. The blebs appeared in crops; as fast as a number had disappeared, new ones appeared, passing through the same stages of development. If the bullæ were punctured with a needle, the fluid escaped, and they collapsed and dried up, looking for a time, however, very much like a flattened variolous pustule.

When I saw the patient, this eruption extended over the entire body, discrete for the most part, but confluent in places. Especially about the genitals was a large patch, very much resembling a patch of confluent smallpox pustules. There was considerable fever present on the day I saw the patient, but very little other evidence of serious constitutional involvement. He was cheerful; strong enough to walk about; slept well, and had a good appetite.

In this case, the diagnosis appeared to me equally plain as in the other. Considering all the symptoms, and not merely the eruption on a

limited portion of the body, which might, and doubtless would, mislead many practitioners who failed to seek further, I gave a diagnosis of pemphigus, and advised the treatment most appropriate, viz., large doses of arsenic.¹

These two cases, especially interesting in view of the consequences involved in an incorrect diagnosis, have led me to direct your attention this evening, as briefly as practicable, to the principal points of difference between pemphigus and such eruptions as might readily be mistaken for it.

Pemphigus is a disease of the skin, characterized by the outbreak of blebs, varying in size from a small bean to a hen's egg, or larger, generally appearing in crops, and accompanied by more or less febrile disturbance.

This definition sufficiently characterizes pemphigus, and marks it as a disease standing by itself. It is not merely an eruption of blebs, but successive crops of these blebs appear. The blebs of pemphigus rise abruptly from the sound skin; have no inflammatory areola, and are, in most cases, tensely filled with a clear, yellowish—sometimes purulent—fluid, or at times containing blood.

In a few days, the fluid is reabsorbed; the roof of the bleb, with some of its contents, dries into a thin scale, which, when removed, leaves a reddened, but otherwise apparently healthy base. If, by means

¹ The patient has entirely recovered under the arsenical treatment, which is claimed by such an accurate observer as Hutchinson to be specific in pemphigus.

of the prick of a needle, or otherwise, the contents of the bleb are discharged, the latter collapses, and dries up, as in the last instance. Unless irritated by mechanical means or stimulating applications, pemphigus blebs rarely contain pus, and no ulceration takes place at their base; hence uncomplicated pemphigus leaves no scars. The blebs consist of single cavities, not subdivided into compartments, as are the pustules and bullæ of smallpox in their earlier stages.

The disease probably most frequently mistaken for pemphigus is impetigo contagiosa. This appears in the form of pea to chestnut-sized blebs, rising abruptly from a non-inflammatory base, but usually flaccid, not tensely filled with fluid like those of pemphigus. The borders of the blebs of impetigo contagiosa are also usually more irregular—not so perfectly rounded or oval as those of pemphigus. They contain a clear fluid, which rapidly becomes changed into a thin, milky pus. The fluid is soon absorbed, or dries with the roof of the bleb into a thin brownish crust with turned-up border lightly adherent at the centre, as if “stuck on” as Tilbury Fox described it. Impetigo contagiosa usually first appears on the face, and, being auto-inoculable, may be transferred to other portions of the body. It is very contagious and usually affects all the children of the same family. In most cases it runs its course in two to four weeks, and hence, probably originate the accounts of epidemics of acute pemphigus, which we so often see in the journals. The resemblance is often very close between the two diseases, and

only a careful investigation will disclose the true nature of the disease in many instances. If the characteristic marks of the two diseases are remembered, however, no mistake should occur.

In a number of cases of erysipelas, frost-bite, burns and scalds, the application of cantharides or mezereon, bullæ appear on the affected part. Here pemphigus can always be excluded by the presence of the uniformly reddened or inflamed base upon which the blebs appear.

In the later stages of acquired syphilis a bullous eruption sometimes appears, which is termed by some authors, "syphilitic pemphigus." The name is misleading, as the eruption of bullæ is the sole point of resemblance. The bullous syphiloderm, as this affection is more properly termed, is differentiated from pemphigus, by an inflammatory areola surrounding the base of the bleb, which becomes purulent, the contents drying into a greenish-brown scab seated upon an ulcerated base, constituting what is called rupia. The bullous syphiloderm is more frequent in children as a manifestation of inherited syphilis.

The early stage of true leprosy is frequently accompanied by an eruption of bullæ. In this disease, however, some hyperæsthesia, followed by anæsthesia of the spots occupied by the blebs generally precedes the eruption. Other concomitant symptoms of grave involvement of the constitution will also be present, and enable the physician to exclude pemphigus.

Smallpox, as the cases before related go to show, may cause a difficulty in diagnosis—a difficulty which is perhaps more serious than that presented

by most other diseases, on account of the results which may ensue if a case of the latter should fail to be recognized. In smallpox, however, the blebs always contain pus or blood ; are not simple cavities, but subdivided into compartments ; are seated upon an inflamed base, and followed by ulceration and loss of substance. The prodromic symptoms of smallpox can also usually be verified in the latter disease ; these do not occur in pemphigus.

In rare cases of exudative erythema, large blebs sometimes occur as one of the multiform manifestations of this disease. The accompanying papules and the generally present patches of diffused red or brownish discoloration will serve to distinguish the affection. Dr. I. E. Atkinson has described some interesting examples of the bullous form of erythema multiforme in *THE MEDICAL NEWS* for December 2, 1882. The so-called herpes iris, which has doubtless sometimes been mistaken for pemphigus, is now generally regarded as merely one of the forms of exudative erythema.

In some rare cases of urticaria, the summit of the wheal is occupied by a bleb, which may simulate the bullous eruption of pemphigus. The presence of other wheals, the urticarial irritability of the skin, and the intense itching in urticaria will serve to distinguish it from pemphigus.

Charcot has pointed out that a bullous eruption sometimes occurs in consequence of nerve-lesions. These eruptions may appear consecutively, simulating the recurrent eruptions in pemphigus. Scars remain in these cases, however, to mark the seat of

the blebs, which is an exceedingly rare result in pemphigus. In the latter disease, also, the eruption would not be so strictly limited to the area supplied by an injured nerve.

Scabies is occasionally accompanied by large bullæ. The presence of papules, pustules, furrows, and excoriations, accompanied by severe itching, and the acarus, discoverable with a lens, would exclude pemphigus.

In ecthyma, large pustules are formed, which may be mistaken for the bullæ of pemphigus. The free pus production, the inflammatory areola around the base of the blebs, and the resulting greenish crusts and superficial ulceration in ecthyma render the diagnosis easy.

An important, possibly frequent, and certainly rarely recognized cause of bullous eruptions is the ingestion of certain medicines. Arsenic, potassium bromide and iodide, quinia, copaiba, and phosphoric acid, have been followed by bullous eruptions, more or less resembling pemphigus. It should in all cases of doubt be ascertained whether such medicines have been taken before deciding upon the diagnosis.

It needs to be added that the practitioner must be constantly on his guard against being victimized by feigned bullous eruptions, *i. e.*, eruptions of blebs caused by the designed application of chemical or dynamical irritants to portions of the skin with intent to deceive. Hysterical women are, of course, the most frequent offenders in this respect, but it must not be forgotten that men sometimes maling

by feigning various formidable skin eruptions. The methods by which bullæ are produced artificially consist in the hot iron, sinapisms, cantharides, strong acids or alkalies, and perhaps in some instances, prolonged pressure. The possibility of this occurring must be constantly borne in mind in order to avoid being discomfited by a malicious or dishonest patient.

While it may be necessary in some cases to defer giving a positive diagnosis for one or several days, close observation, and recollection of the points of distinction here laid down will nearly always enable the practitioner to differentiate between pemphigus and other bullous eruptions.

